

OBJECTIVES

To determine the National incidence and prevalence of dementia diagnosed in children under the age of fourteen; to identify the aetiological profile of childhood dementia; to determine the geographical distribution of children suffering from this condition and; to assess the psychosocial and neuropsychiatric needs of this population.

INVESTIGATOR CONTACT DETAILS (*Principal Investigator)

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SUMMARY PROTOCOL

Dementia in childhood has been thought to be quite rare. Many of the children have a common clinical picture of multiple developmental (including cognitive) skill loss. Despite this common clinical picture they have diverse aetiologies. However, the incidence and prevalence of children with dementia has never been established in any study to date.

CASE DEFINITION

Any child who suffers from an illness fulfilling the following criteria:

1. **Multiple losses of already attained developmental skills**
These might include the loss of visual tracking and following in infants, the loss of walking or speech in toddlers, or deterioration in school performance, eg., marked deterioration in writing ability in older children and memory loss with loss of other cognitive skills in adolescence.
2. **Duration of illness greater than 3 months**
3. **Skill loss most probably due to CNS dysfunction**
Association with seizures, loss of vision or delirium all suggest CNS dysfunction
4. **Evidence of generalised brain dysfunction (not merely focal)**
eg., specific acquired language impairment or a seizure disorder alone would be insufficient unless there was evidence of global deterioration and dysfunction.
5. **The condition must not be explicable in terms of acute drug toxicity, prolonged ictal confusion or other causes of delirium**

Examples of aetiologies associated with childhood dementia include:

- Degenerative** – eg., Batten’s disease, Adrenoleucodystrophy, SSPE, mitochondrial cytopathies
- Infectious** – eg., HIV Dementia, post-encephalitis
- Traumatic** – eg., secondary to head injuries
- Anoxic** – eg., secondary to drowning

REPORTING INSTRUCTIONS

Please report any **new** or **old** patients **under 14 year of age** with dementia seen in the last month who you have not previously reported.

FOLLOW-UP OF POSITIVE RETURNS

A questionnaire requesting further details will be forwarded to the practitioners who report a case.